CASE OF SPORADIC CRETINISM TREATED WITH THYROID GLAND.

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CASE OF SPORADIC CRETINISM TREATED WITH THYROID GLAND.

In the British Medical Journal of March 28th, 1891, Dr. T. C. Railton, of Manchester, published an account of two sporadic cretins, who were brothers, and gave their portraits.

The younger of the brothers — D. B. —was admitted to the Royal Albert Asylum, Lancaster, on August 5th, 1891, and has been here since that date. On March 27th, 1893, at the suggestion of Dr. Shuttleworth, I commenced to treat the patient by thyroid feeding, so that the full history of the case may be of interest.

Family History of Patient.

His father, aged 45, is a fish salesman in Manchester. He is temperate, and is an intelligent man. He is stout and measures about 5 feet 6 inches in height. Is subject to asthma, as was his (the father's) mother. There is no history of intemperance in his family. The patient's mother is aged 35. She is temperate, and an intelligent, but very neurotic, woman, and inclined to be delicate. Her father was intemperate, and died at the age of 44 of phthisis. There is no further history of intemperance in her family. The father and mother of the patient were not related, and there is no history of consanguineous marriages in their families. There is no history of insanity on either side.

History of Patient.

The patient was born on June 24th, 1884. He is the fourth born child. His eldest brother, aged 14, is also a cretin. A healthy brother and sister were born between the patient and his eldest brother. There are five brothers and sisters healthy and normal, physically and mentally. D. B. was born at full time and normally; labour was protracted. No instruments were used, and he was not asphyxiated when born. He was not convulsed soon after birth, nor has he ever had any kind of fit. He is said to have been bright as an infant, and cut his first teeth early. His mental deficiency began to be noticed at about 2 years of age. He did not begin to walk till about 2½ years. He has had measles, whooping cough, and diphtheria.

G. B., 11 years

D. B., 6 years 3 months.

Present condition of G. B. and D. B.

Description of Patient on Admission to the Royal Albert Asylum
(August 5th, 1891)

A well-marked case of sporadic cretinism. He has a rather pleasant expression and bright smile. His temperament is phlegmatic, and he is slow and deliberate in all his movements, even a smile taking a long time to spread over his face. He seems to take some interest in his surroundings, but will not speak. His mouth is always open and his teeth slightly parted, showing the extremity of his tongue, which is thick and very blunt, and rounded at the end; it seemed somewhat too large for the mouth. The papillæ on the tongue are small, and the surface is smooth and rather pale on the upper surface; on the lower it tends to be blue. His palate is normal in height and formation. He still has his first set of teeth, which are set widely apart; the molars are carious. He does not slaver. His mouth is wide, but the lips are fairly well formed and of a good colour; they are not everted, but the lower is larger than normal. His nose is wide, especially at the root, and the septum and alæ are thick; the nose is short and rather turned up; the nostrils are broad. His eyes are bright and his eyelids are well formed and horizontal, as are also the eyebrows, which, with the eyelashes, are of normal thickness and length, and are well placed. Pupils medium in size and react well to light and accommodation. Ears well formed and normal in size and position. His skin is rather dry and rough on the face, but on the unexposed surface of his body it is smooth, though dry. His complexion is pale, with a tinge of sallowness, and a rather waxy look. Malar blush very slight. His head is large. Circumference 21 inches. Dolichocephalic. Cephalic index 73.3. Fontanelles closed. There is a distinct ridge to be felt over the sutures on the vertex. His hair is rather thin, straight and dry, and grows slowly. He has a large and protuberant abdomen, and a small umbilical hernia. Chest circular and barrel-shaped. His legs are bowed, and the ankles are enlarged. Feet short and square. Second toe small, and over-riding third. Hand square and short. Good grasp. Wrists enlarged. Slight beading of ribs. Respiration normal. Heart sounds weak, with a faint systolic murmur at apex. He walks slowly, and in a heavy manner. His run is a very awkward waddle. Kneejerk slightly exaggerated. Skin sensibility normal. Extremities cold. A small portion of thyroid can be felt. No pseudo-lipomata. No exaggeration of the spinal curves. His bowels are constipated. He is easily teased or frightened, and if his temper is ruffled he has prolonged fits of sulkiness, during which he will neither smile nor show any sign of friendliness. He was over a month in the asylum before he was heard to utter a word.

Effects of Treatment.

Thyroid treatment was commenced on March 27th, 1893. He was given not quite a quarter of one lobe of a fresh sheep's thyroid minced, and mixed with some warm rice and jam at tea-time. He took it well without perceiving any unpleasant taste.

March 28th. He complained of headache, vomited several times and took no breakfast. Looked very depressed and was very pale and cold. Put to bed.

March 29th. The vomiting continued in the early morning, but he took a little limewater and milk later on. The tongue was clean. The bowels had not been moved since March 27th. He was very depressed and apathetic, and complained of headache. He did not vomit again, however, and the headache was better in the evening. He was kept on a diet of beef-tea and milk, and was ordered calomel, gr. ij, at bedtime.

March 30th. The bowels acted; no headache nor sickness; he took breakfast of bread and milk, was bright and smiled. On April 1st he was quite himself again.

April 13th. He was ordered ½ part of the fresh lobe twice a week, minced in rice and jam. He took it well and showed no unpleasant after-effects.

April 26th. He was brighter and more active, spoke much more readily, and had become playful and lively. His face was beginning to lose the cretinoid appearance, and the fea-

tures were all sharper and clearer. His skin was becoming softer and smoother. He had had a kind of scurfy, dirty patch on his nose and forehead, which soap would not remove; this came off almost in one piece, like a crust. He perspired rather copiously, especially about the head.

May 13th. The skin on the feet and hands was peeling. He was much brighter and more active. His tongue was now normal in size and appearance, having lost the thick blunt shape and blue tinge. He could run fairly fast without awkwardness.

July 6th. Mentally he was wonderfully bright; he liked to talk and answer questions, and had learnt "Little Jack Horner" from a girl patient. His vocabulary was enlarging considerably. He was full of spontaneous playfulness and mischief. Physically he had got very thin, except in the face, which was plump and healthy-looking without the least sign of puffiness. His appetite was good, and his bowels regular, without constipation. He was cutting two lower central incisors.

On June 5th the dose was diminished to one twelfth, as he was getting thin.

He did not take the gland after July 7th, and, on September 13th, it was noted that he began to show signs of slowly reverting to his former state. The condition of solid œdema was quite perceptible again. His speech was not so distinct nor so quick, owing to the tongue beginning to thicken and to his mental state not being so active. He was not so bright, and he had lost some of his spontaneity. On September 16th he was put on one tabloid (Burroughs and Wellcome) once every day at dinner time—equivalent to one-twelfth of a lobe. On October 18th I took him to Manchester to go home to his parents for a holiday, and showed him that evening at the Manchester Medical Society. (Dr. Railton also showing the eldest brother, whom he was treating with thyroid.) The members of the Society, who had seen the two brothers before, were struck by the very marked change for the better in them both.

December 13th. The patient returned to the Royal Albert Asylum from home, where he had continued taking one tabloid every day. His mother said he had become so active and mischievous that she could hardly put up with him. His father's report, on bringing him back, was: "I find that my son, D. B., has improved very much; he is much brighter and more cheerful and intelligent than before in every way." He was now cutting two upper central incisors.

February 16th, 1894 He has continued to take one tabloid a day, and maintains his mental and physical improvement. He is growing taller, and is active in all his functions. There are at present few traces of sporadic cretinism about him. The umbilical hernia is now imperceptible. His average temperature before treatment was 95°; since treatment it has been about 98°.

Remarks.

Dr. Byrom Bramwell, in his "Clinical Remarks on a Case of Sporadic Cretinism," in the British Medical Journal of January 6th, notices that "an umbilical hernia is nearly always present in cases of sporadic cretinism;" this is certainly so in the greater number of the cases, but it is perhaps worth recording that there have been two (out of four) rather

extreme cases of cretinism in the Royal Albert Asylum in which this deformity did not exist—both girls.

It is also remarkable that, where the hernia has been small, improvement and even disappearance of the hernia has taken place during the thyroid treatment, as occurred in the boy D. B., and in the case published by Dr. John Thomson in the *Edinburgh Medical Journal* for February 1894, and in cases published by Dr. A. Gordon Paterson and by Dr. John B. Hellier in the *Lancet* of November 4th, 1893.

The constancy with which numerous symptoms which accompany rickets also occur in cases of sporadic cretinism is noticeable. Thus we almost invariably find the tibiæ bent, and the ankles and wrists enlarged, while beading of the ribs is sometimes present, and altered or exaggerated spinal curves. Also the open fontanelles, barrel-shaped chest, and protuberant abdomen, delayed dentition, and lateness in walking and talking. In the boy, D. B., there is decided thickening of the cranial bones along their edges, some of the sutures feeling like ridges. The absence of perspiration in sporadic cretinism is very constant. However, the boy D. B., since thyroid treatment was commenced, sweats remarkably copiously when asleep, especially about the head and neck.

Table of Measurements.

Date.	Height	Weight	Circumference round Navel.	Circumference round Mammæ.	Circumference round Calf.
1891.	Ins.	lbs.	Ins.	Ins.	Ins.
March ...	33	32½		—	—
August ...	34	35	—	—	—
1893.					
March ...	34¾	40	22	21½	7¾
April ...	—	38	20	21	—
May ...	—	37½	19	20¾	—
June ...	—	37½	19	21	7½
August ...	37	43	22½	21½	8
September	—	43½	22	21¼	8¼
October ...	—	40	21	21½	8
November	—	38½	—	21½	—
1894.					
January ...	39	41½	22	22	8
February...	—	—	22½	22	8
April 1899	51½	66¼	23½	25½	9 7/8

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Head Measurements.

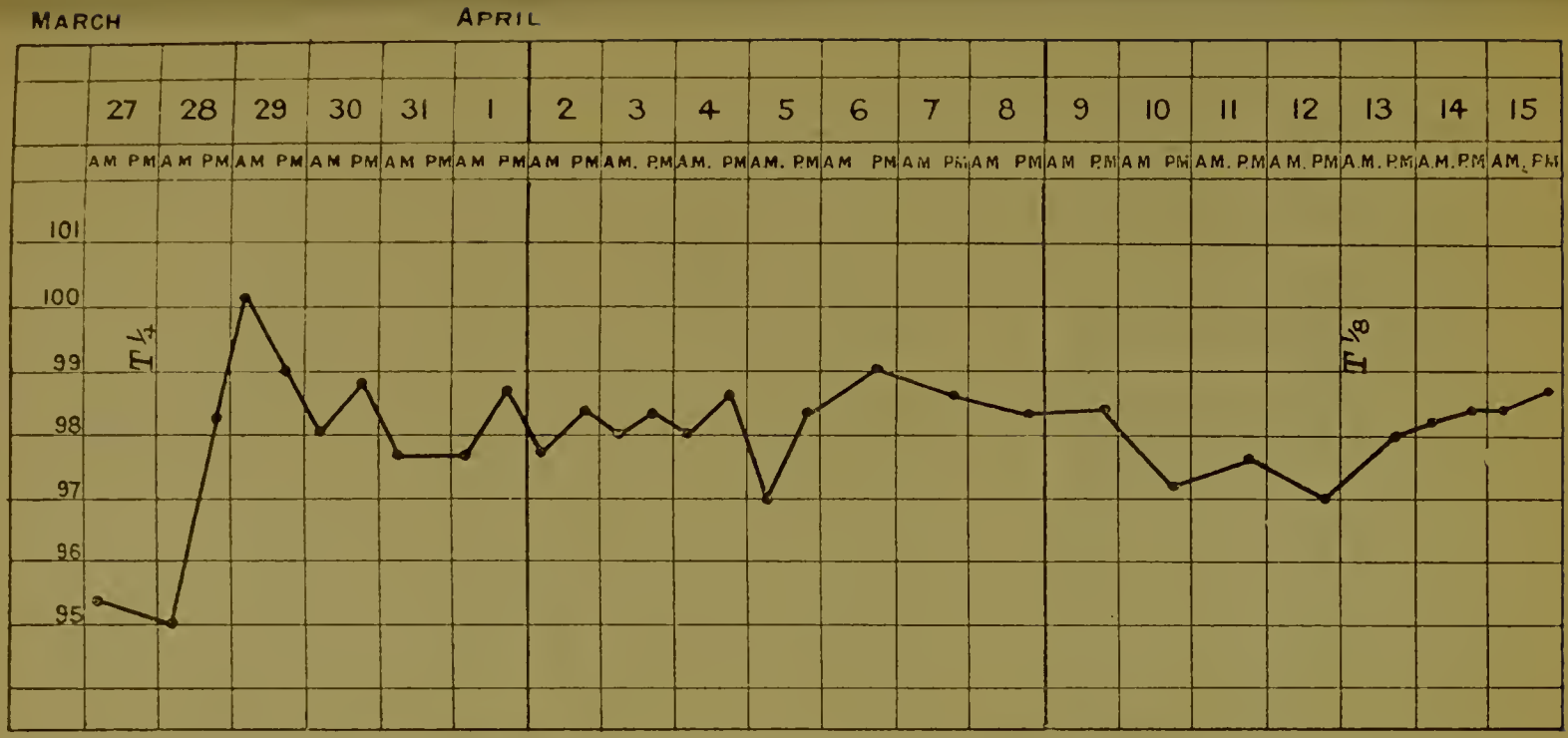
22 Circumference 21 ins. ... Above ears and over occipital tuberosity.

(a) 14¼ Transverse (*a*) 13 ins., (*b*) 4 ins. ... ... { (*a*) Tape measure from ear to ear over vertex. (*b*) Calliper measure from ear to ear over vertex.

(a) 13 7/8 Longitudinal (*a*) 13 ins , (*b*) 7½ ins. ... ... { (*a*) Tape measure from nasal notch to occipital tuberosity. (*b*) Calliper measure from nasal notch to occipital tuberosity.

5 5/8 Width of forehead 4¾ ins. ... { Between external angular processes of frontal.

In investigating the etiology of sporadic cretinism, it is striking how prominently "maternal depression and worry during pregnancy" seem to stand out among the alleged causes. In the case of both D. B. and his brother, the mother (a neurotic woman) alleges abnormal depression while pregnant with each of them (and not during the pregnancies



Temperature chart, case of D. B. at the commencement of treatment, showing the rise of temperature which followed the administration of a quarter of a lobe of fresh sheep's thyroid on March 27th, the maintenance of a relatively high temperature (as compared with the temperature of 95° to 96° before treatment) for 11 days, a decline, and a second rise of temperature after the administration of one-eighth of a lobe on April 13th.

of the other normal children). In the case of another sporadic cretin in the Royal Albert Asylum at present, the most prominent cause to be found in the family history is unusual and great worry and depression on the part of the mother during her pregnancy with the patient (owing to money difficulties), all her other children being remarkably fine specimens, as is she herself and her husband. May it not be that the atrophic condition of the thyroid gland, which exists in sporadic cretinism, is brought about by a numerous class of causes, all of which tend to produce slow impairment of nutrition in the fœtus ?

Among the more prominent of these causes we might expect to find maternal depression and worry, or a lowered vitality in the parents produced by bad air or food, cold damp houses, or insufficient sunlight and want of cleanliness.

In the family histories of the patients it is remarkable that goître, consanguinity, insanity, intemperance, phthisis, or syphilis seldom appear to be present.